CASE REPORT

C1 fracture in a patient with a congenital cleft in the posterior arch: report on a failed conservative treatment

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SUMMARY

Unstable burst fractures of the atlas require sufficient immobilisation either with an external device or by surgical fusion. In patients with pre-existing congenital atlantal defects the decision whether to recommend non-operative or operative therapy is difficult. Treatment options are controversially discussed and standardised guidelines are not available. Although most surgeons advise operative fusion or osteosynthesis in atlanto-axial instability, non-operative immobilisation can be successful. We report on a patient with an unstable C1 burst fracture with a congenital posterior cleft in the atlantal arch who was treated with cervical extension and halo-vest immobilisation. Although callus bridging was initially observed, 9 months after the injury there was no sufficient fracture consolidation and surgical fusion had to be advised. While non-operative treatment may work in patients without congenital defects of the C1 arch, such is not recommended in patients who present with this anomaly.

BACKGROUND

Atlas fractures are very common in cervical spine trauma and account for 25% of craniocervical injuries with widely accepted treatment guidelines.¹⁻⁴ The spine surgeon may, however, encounter problems in selecting the treatment strategy when a patient is found to have a congenital bony anomaly. Because these defects are rare, there is no guideline on how to treat an atlas fracture when such defect is present. The atlanto-axial stability can be compromised which also renders these patients at greater fracture risk.⁵⁻⁶ It is unclear whether this subgroup should be treated by operative stabilisation or by immobilisation. There is only one reported case in literature⁷ where an atlas fracture has been treated in a patient with a posterior cleft. In this patient, the fracture healed after 2 months of cast immobilisation.⁷ It was however not reported whether the transverse ligament was intact.⁷

Congenital anomalies of the atlas appear as clefts in the arch varying in size and location.⁸ Posterior defects are more common (4%) than anterior (0.1%) ones.⁸⁻⁹ Most of them are found incidentally and clinical symptoms are infrequent.⁸ When a mobile, unfused isolated posterior arch tubercle is present, patients may develop chronic neurological symptoms or acute deficits following trauma.⁸⁻¹⁰ The key element for deciding whether to opt for surgical stabilisation or conservative treatment in

cases of an isolated fracture is the integrity of the transverse ligament.¹¹ It is crucial for the stability of the atlanto-axial junction.¹¹⁻¹² When intact, good outcomes were reported for halo-brace immobilisation.¹³ When torn in its substance, surgical fusion is typically enforced.¹¹ When avulsed from the lateral mass, non-surgical treatment is optional with reportedly successful outcome.¹⁴ We report on a patient with a congenital cleft in the posterior atlantal arch that presented with a combined, unstable C1 burst fracture. The transverse ligament was avulsed from the lateral mass and the patient was initially treated with a Gardner-Wells extension device for 2 weeks and consecutive halo immobilisation for another 2 months.

CASE PRESENTATION

A 26-year-old man was assigned to our department with the diagnosis of a C1 fracture after he had jumped into a wave during swimming in the sea and hit the ground with his forehead. The patient was of very good overall physique and very high compliance. On the initial radiograph, the fracture had been missed and the patient was sent home. On re-evaluation of the radiographs, a posterior arch defect was observed. A subsequent CT-scan revealed a left anterior arch fracture of the atlas (Gehweiler Type II)¹ and a congenital cleft of the posterior atlas arch (Currarino Type A)⁸ (figure 1). An osseous fragment next to the right cranial portion of the odontoid process indicated an avulsion of the transverse ligament from the lateral mass (Dickman Type II).¹¹⁻¹² MR images were obtained in order to assess further ligament injury. On MRI, the transverse ligament was found intact in its ligamentous substance. The anterior and posterior longitudinal ligaments, the interspinous and flava ligaments as well as the intervertebral disks were found intact. There was no indication of spinal canal stenosis or damage to the spinal cord. The atlanto-axial interval (ADI) was 2.5 mm and the lateral mass displacement was <6.9 mm on odontoid view radiographs. On presentation the patient had no neurological deficits and reported tolerable pain under oral analgesics. He never experienced neurological deficits or cervical pain before the accident. The treatment strategy was discussed with the patient and conservative treatment was chosen. He was sent home and immobilised in a cervical collar. After 1 week, the radiographical follow-up showed an increase of the atlanto-axial distance (2.5 → 5 mm) and an increase of lateral

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Figure 1 Post-traumatic CT scan showing the congenital posterior arch defect (A) (type A according to Curriano⁸) and an anterior arch fracture (B) (type II according to Gehweiler¹). A small avulsed fragment (C) on the right portion of the odontoid process indicates a transverse ligament avulsion (type II according to Dickman¹¹).

mass displacement (<6.9 mm→9 mm). Fracture instability was confirmed under dynamic fluoroscopy under cervical flexion and extension. The treatment options were discussed with the patient and operative fusion was proposed. Two methods, a posterior C1–C2 fusion according to Harms¹⁵ or a transoral C1 osteosynthesis, according to Schmelzle and Harms,¹⁶ were suggested. The patient refused an operation due to loss of cervical mobility in posterior C1–C2 fusion and due to the risk for infection in anterior C1 reduction and internal fixation. Alternatively, cervical extension therapy in a Gardner-Wells clamp and adjacent halo-vest stabilisation were chosen.

Hence, we administered an extension therapy with an initial onset of a Gardner-Wells extension (7 kg/15.4 lbs) for 2 weeks (figure 2) and a halo-vest extension for another 8 weeks (figure 3). The halo-vest extension was readjusted under fluoroscopy after 2, 3, 13 and 17 days to assure a sufficient reposition. The halo vest was removed after 57 days (8 weeks). Bridging callus was

documented on CT (figure 4). Four weeks after removal of the halo vest the patient was pain-free and lateral mass displacement was <6.9 mm on an open-mouth odontoid view (3.1 mm left, 2.0 mm right). The ADI on a lateral radiograph was 3.6 mm. A physiotherapy protocol with careful passive mobilisation was initiated in order to improve upper spine mobility. According to the protocol, the treatment was defined as successful.

OUTCOME AND FOLLOW-UP

However, 4 months later, at a total of 9 months after the injury, a fracture gap was observed with a distance of 13 mm (figure 5). The ADI was 4 mm and lateral mass displacement was 10 mm (5 mm on both sides). Therefore, surgical stabilisation with posterior fusion was recommended to the patient. He still refused an operative treatment and decided for further conservative treatment.

DISCUSSION

The treatment strategy for C1 fractures in patients with congenital defects of the atlantal ring is challenging for the surgeon and the patient. In the presented case, a conservative strategy failed although a bony interosseous bridge was observed at the end of the immobilisation period.

Posterior atlantal arch defects are attributed to the defective or absent development of the cartilaginous preformation of the arch.¹⁷ They are considered benign anatomical variations and usually found incidentally.⁸ Autopsies and surgical explorations have shown that on the one hand dense fibrous bands that bridge the bony gaps may account for a good stability. A membrane extending from the posterior margin of the foramen magnum to the superior border of the axis was also interpreted to account for sufficient cervical stability.⁸ On the other hand, these clefts were associated to atlanto-axial instability and neurological deficits.^{18 19}

Atlantal fractures are very common in cervical spine injury. This is a result of the medio-caudal inclination of the articular facets, which transmit the axial loading force of the occiput and exert a radial force on the lateral masses, the atlantal ring

Figure 2 Extension therapy in a bed with a Gardner-Wells clamp extending the cervical spine with 7 kg (=15.4 lbs). The bed was inclined 10° in order to eliminate cranial gliding of the patient on the mattress.

Figure 3 Halo-vest extension was conducted for 8 weeks after initial extension in a Gardner-Wells clamp. After 2, 3, 13 and 17 days the vest was readjusted under fluoroscopic control to achieve sufficient reposition.

expands and most often fractures at its weakest points where the anterior and posterior arches attach to the lateral masses. The first fracture classification was presented by Jefferson in 1920²⁰ which was later modified by Landells and van Petegham²¹ and by Gehweiler.¹ C1/C2 biomechanics are hypothesised to be altered when the posterior arch is not fused which may predispose for a higher injury risk which was not yet shown by greater incidence in literature.¹⁰ There is only one case⁷ in which an anterior arch fracture has been reported in association with a posterior atlantal cleft. This patient was successfully treated by immobilisation in a cast for 2 months.

Figure 4 Three-dimensional-CT image reconstructions before the halo-vest was removed after 57 days (8 weeks). The bony bridge (arrow) in the fracture zone of the anterior arch evidenced bony healing.

Figure 5 CT of the atlas 9 months after the injury. There is a fracture gap (B) without osseous healing. The bony bridge observed prior to halo-vest removal is not detectable.

However, these authors do not describe the integrity of the transverse ligament which is a crucial stabiliser.⁷ There are no data in literature associating congenital clefts with a higher fracture risk. This lack of data may indirectly indicate no such increased risk. However, a low prevalence of congenital anomalies has to be considered. Further biomechanical or epidemiological data are needed to address this issue.

In the present case different classifications were needed to describe the fracture: It was summarised as a C3.2 (AO classification) combination injury, according to Aebi and Nazarian²² including:

1. Gehweiler type II¹ fracture of the anterior arch.
2. Type A cleft of the posterior arch according to Cuarrino *et al.*⁸
3. Type II transverse ligament injury according to Dickman *et al.*¹¹

To evaluate C1 fractures using plain radiographs, it is essential to obtain lateral and open-mouth odontoid views. A thin slice CT scan is mandatory for optimal fracture classification.²³ But the most critical aspect for therapy decision is to determine the stability of the atlanto-axial junction. It is maintained by the transverse ligament and best assessed on high-resolution MRI.¹¹

Radiographs may lead the physician to obtain CT and MRI when the ADI on lateral radiographs is >3 mm in adults and >5 mm in children.¹³ They are not sensitive enough and not recommended for therapy decision. The 'Rule of Spence', assumes instability when the sum of lateral masses displacement exceeds 6.9 mm on an open-mouth odontoid view.²⁴ Dickman *et al.*¹¹ reported 61% of missed ruptures when applying this rule.

Regardless of whether C1 fractures occur in isolation or with other cervical spine fractures, no standards or guidelines have been established²⁵ and recommendations for the treatment of fractures in patients with congenital clefts are not available. Cervical traction can align the splayed lateral masses through ligamentotaxis and unstable atlantal burst fractures may therefore heal satisfactorily with immobilisation in halo traction or halo vest without surgery.^{3 4 26 27} A complete reposition is however not always achieved and the functional outcome of a healed but malpositioned fracture is poor.²⁸

With surgery, adequate reposition and stability are well documented. The disadvantage of a fusion is the elimination of rotation at the C1–C2 junction. Communicative movements, for example, turning of the head, predominantly occur in this joint.²³ When fused, the more distal cervical vertebrae have to compensate and consequently, the range of motion is reduced simultaneously increasing the degeneration of the lower cervical

spine.²⁹ In a transarticular fusion described by Magerl *et al*³⁰ only the anteroflexion and retroflexion in C0/C1 is preserved. The main limitation relates to the anatomic variations of the vertebral artery precluding safe screw placement.³¹ The injury risk was reported from 4% to 14%.³¹ In 20% of specimen in a cadaveric study a bilateral screw placement was impossible due to anatomic variations that placed the vertebral artery at risk.^{32–33} In response to these limitations new techniques evolved with independently placed C1 and C2 screws connected with rods or plates. Satisfactory screw placements and solid fusions after 1 year were reported.^{15–31}

In order to preserve the atlantoaxial rotation Schmelzle and Harms¹⁶ offered a transoral approach where isolated reduction and osteosynthesis of C1 preserves a total average range of motion in C1–C2 of 39.2°. This is a substantial amount of mobility, but still less than that found in a healthy population or in cadaver studies.^{34–35} However, the transoral operation has relative disadvantages, including high risk of postoperative infection and a very high level of surgical experience needed.^{36–37}

The decision of whether to pursue surgical intervention is dependent on the type of transverse ligament injury. Dickman *et al*¹¹ classified two main types which imply a different treatment. Type I injuries are intrasubstance ligament ruptures and incapable to heal without surgical immobilisation. Type II injuries represent avulsions of the transverse ligament on the C1 lateral mass that can heal if the fracture is immobilised non-operatively.¹⁴ Therefore, considerable lateral mass widening may not necessarily be accompanied with instability.¹⁴ This is because residual stability arises from the alar ligaments and facet capsules which render non-surgical therapy as a viable treatment option.¹⁴

This however may not be true for patients with a congenital posterior cleft in the atlantal arch. In these patients, the residual stability may not suffice for conservative treatment of a transverse ligament avulsion. Although an adequate healing process was documented in this patient after 8 weeks of halo immobilisation the overall bone proliferation was insufficient on the long run. This could be interpreted in the light of higher micro-motions occurring at the fracture site, which might be promoted by the posterior cleft. The fibrous bands bridging the gap may not suffice the necessities of rigidity to account for bony healing. A Gardner-Wells extension for 2 weeks and halo-vest immobilisation for 8 weeks including subsequent follow-up consultations with vest adjustment under fluoroscopic control, and a highly compliant patient were incapable to allow for transverse ligament scarring and bone healing.

Learning points

- ▶ Halo immobilisation following cervical extension is not recommended in patients with an unstable C1 burst fracture and transverse ligament avulsion who have a congenital cleft in the posterior arch.
- ▶ A congenital posterior arch defect may adversely affect sufficient fracture healing in C1 burst fractures.
- ▶ If non-surgical treatment is inevitable, a halo immobilisation period of more than 8 weeks should be considered.

Competing interests None.

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